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CASE REPORT

Aggressive chondroblastic osteosarcoma of the jawbone

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Summary Osteosarcoma of the craniofacial bones (CFOS) usually behaves less aggressively than appendicular skeleton OS, resulting in a better therapeutic response to either surgery alone or multi-modality treatment. Here we report the case of a 54-year old male diagnosed with high-grade chondroblastic OS of the mandible, who presented with painful swelling of the horizontal branch of the right mandible, accompanied by paresthesias and paroxystic pain episodes. The patient was treated with aggressive surgery and adjuvant chemotherapy but, despite favorable clinical prognostic factors, disease rapidly progressed to local and distant relapse, causing the patient's death 18 months after diagnosis.

Introduction

Compared to appendicular skeleton osteosarcoma, osteosarcoma of the craniofacial bones (CFOS) has better prognosis and rarely develops distant metastases.[1–3] Here we report a case of unusually aggressive, high-grade, chondroblastic OS of the mandible.

Case report

R.L., a 54-year old male, presented in March 2003 with a 3-month history of painful swelling of the horizontal branch of the right mandible, accompanied by paresthesias and paroxystic pain episodes in the right mandibular trigeminal branch distribution territory. Orthopantomographic exam, inferior occlusal X-rays, and dental CT scan demonstrated limited interradicular osteolysis of 4.6, with reduction of bone density in the right horizontal branch (Fig. 1A–C). Contrast-enhanced MRI revealed

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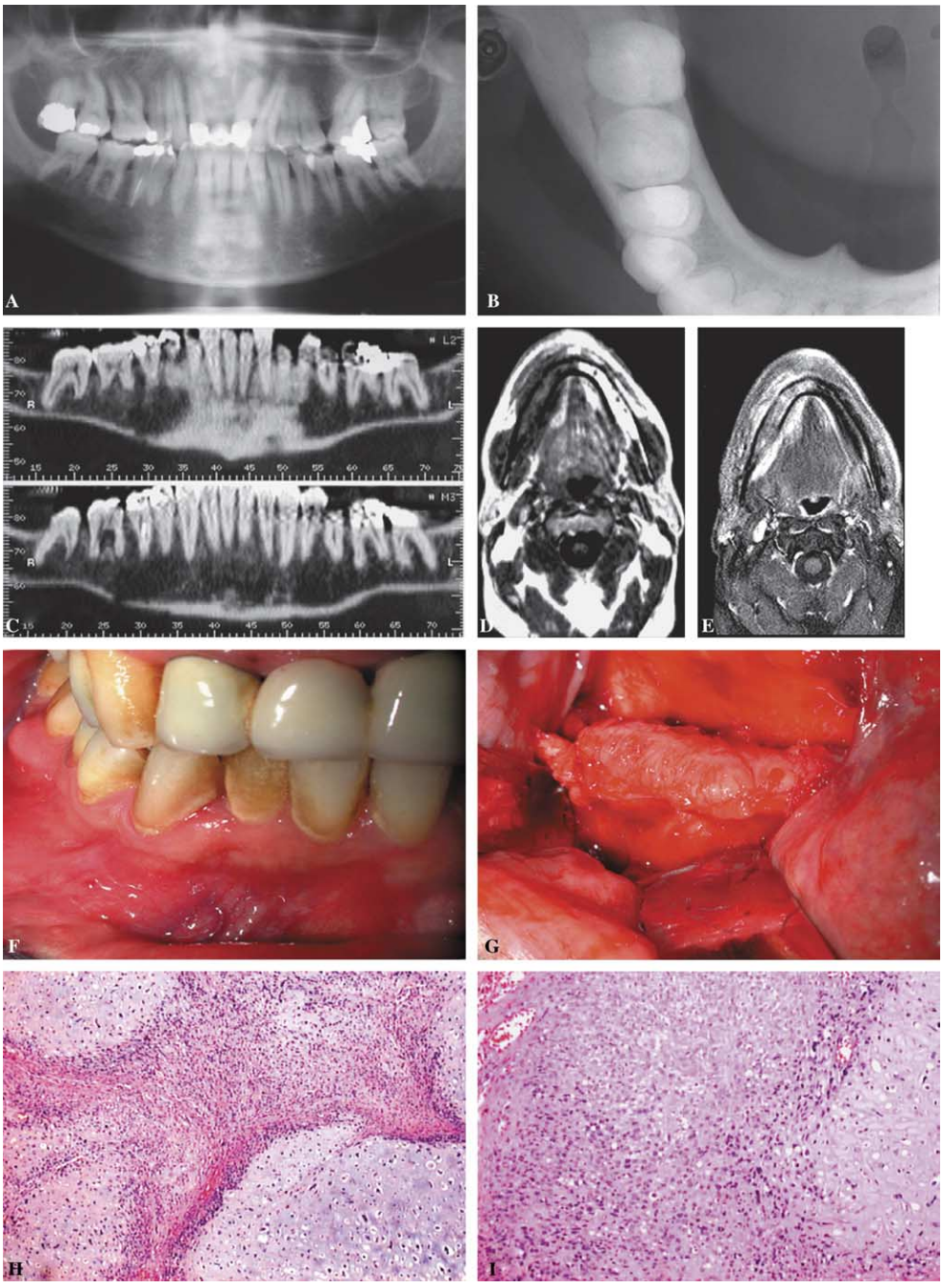

Figure 1 (A) Orthopantomographic exam revealing an area of osteolysis in the right mandibular body, particularly in the periapical region of 4.6, with shedding of the mesial root of 4.7. (B) Inferior occlusal X-rays showing apical and periodontal reduction of bone mineral density of 4.5 and 4.4 involving the right emimandibula and the roots of 4.6 and 4.7. (C) Dental CT Scan demonstrated limited interradicular osteolysis of 4.6, with reduction of bone density in the right horizontal branch. (D) and (E) Post contrast T1W STIR and T2 MRI demonstrating the extent of bone and soft tissue involvement. The lesion appears hypointense in T1 and hyperintense in T2 and shows contrast-enhancement. (F) Intraoral examination revealing a normal mucosa with congested areas. (G) Surgical examination showing a white-gray, translucent mass, adhering loosely to the bone and firmly to the periostium. (H) High grade chondroblastic OS; chondroblastic activity with atypical osteosarcoma cells arranged in palisades and sometimes in vortexes (EE 10X). (I) At higher magnification (EE 20X) small atypical osteoblasts with one or more nuclei in the contest of dilated blood vessels and polymorphic chondroblastic proliferation are observed.

extensive alteration of the horizontal branch of the right half of the mandible from the angle to the symphisis with partial involvement of the vertical branch up to the sigmoid incisure and extension to the surrounding soft tissues (Fig. 1D—E). Biopsy (Fig. 1F—G) revealed small, atypical, sometimes multi-nucleated osteoblasts in the context of dilated blood vessels and polymorphic chondroblastic proliferation, allowing a diagnosis of high-grade chondroblastic OS (Fig. 1H—I). Right emimandibulectomy followed by reconstruction with fibular osteocutaneous free flap was performed. Histological examination confirmed the diagnosis of high-grade OS of the mandible and demonstrated complete excision of a 8 cm lesion, with wide (>5 mm) margins. During adjuvant polychemotherapy, the patient experienced local relapse and underwent salvage surgery (total mandibulectomy) followed by external beam radiation therapy, but died of further local relapse and pulmonary metastatic disease 18 months after diagnosis, despite palliative chemotherapy.

Discussion

CFOS accounts for 6—10% of all OS, with mandibular location in approximately 40% of patients.[1—3] Several differences in disease characteristics distinguish CFOS and OS at other primary sites: CFOS most commonly affects patients in their 30 s; distant metastases are rarely reported (10—20% of patients); survival after surgery alone has historically been better for CFOS than for OS arising at other locations, with reported 5-year survival rates of 23—37% and 10—20% for CFOS and other OS, respectively.[1—6]

Complete surgical excision with adequate margins (>5 mm) is the mainstay of treatment for CFOS.[1—6] Conversely, the role of pre-and/or post-operative chemotherapy is more controversial. Although two recent meta-analysis have yielded conflicting results[4,5] and despite the lack in survival differences between patients treated with surgery alone and surgery + chemotherapy in the largest series reported to date,[1] the prognosis of CFOS has steadily improved over the past 20 years, at least in part due the introduction of multi-agent chemotherapy.

Factors associated with a poor prognosis are age older than 60 years, non-mandibular location, tumor size >6 cm, osteoblastic histology, advanced stage, non-surgical initial therapy, and positive resection margins.[1] In the case of mandibular OS presented here, despite the presence of tumor size >6 cm as the only adverse prognostic factor, disease displayed an extremely aggressive clinical course, with local relapse occurring during adjuvant chemotherapy and development of distant metastases early in the course of disease. Such an unusual behavior highlights the inadequacy of clinical parameters to correctly predict prognosis in individual patients and the need for a better understanding of the biology of CFOS, as well as of other malignancies.

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